

when it involves the skin and other systems, most commonly the gastrointestinal tract and the central nervous system. The malignant type is most common and is typically fatal within two to three years from the time of onset of systemic involvement. The most common complications and cause of death for subjects with Degos Disease are gastrointestinal tract complications. The less common variant is the benign type with only cutaneous involvement. In the skin, it manifests with erythematous, pink or red papules. After healing, these papules leave scars with pathognomonic, central, porcelain white atrophic centers and a peripheral telangiectatic rim.

In a case by Pierce and Smith, they mention chronic pleuritis and pericarditis were the major causes of morbidity. There are very few reported cases of pleuritis and pericarditis, most diagnosed as incidental findings at autopsy. Another case by Notash et al. mentioned that severe restrictive cardiopulmonary insufficiency led to death of a patient with Degos disease. Another case with cardiopulmonary involvement was reported in 1996. An infant case of Degos disease in which the patient died due to disseminated myocardial infarct was presented by Cabre et al. Also, two cases of constrictive pericarditis have been described. One of these cases underwent a pericardiectomy but died four months later due to an intestinal hemorrhage. The other case, described by Pierce and Smith, died due to heart failure and respiratory disorder three months after a left lung decortication. Histology of the heart of both cases indicated myocardial impairment. Pierce and Smith suggested that the myocardial impairment was present with no evidence of associated lesions. Pleuritis and pericarditis were reported as symptoms of Degos Disease by Voigt et al. One case with calcific constrictive pericarditis and recurrent pleural effusions in a patient with long-standing Degos Disease was also reported. Another case by Shahshahani et al presented a case in which the patient died due to severe cardiopulmonary failure and the autopsy revealed severe diffuse fibrotic thickening of the pericardium and pleura. One case with endocardial involvement and positive anticardiolipin antibodies has been reported. Malignant atrophic papulosis (Degos' disease) was first described by Kohlmeier in 1941 and recognized as a specific entity by Degos in 1942.

Cardiopulmonary involvement is a rare but serious complication of DD. These complications must be considered in the disease spectrum for this form of vasculopathy. Our case is unique as it was diagnosed in a living patient currently being treated with medications. The medications he was prescribed were cyclosporine, eculizumab, and enoxaparin sodium. Had the illness not been diagnosed at an early stage, there is a possibility the patient would not have survived. In this case, the pericarditis appeared to be triggered by endothelial cell injury with resultant thrombosis and supervening ischemic complications. There is limited literature on this topic.

[Interventional Management]

Procedural step:

N/A

Case Summary:

Cardiopulmonary involvement is a rare but serious complication of DD. These complications must be considered in the disease spectrum for this form of vasculopathy. Our case is unique as it was diagnosed in a living patient currently being treated with medications. The medications he was prescribed were cyclosporine, eculizumab, and enoxaparin sodium. Had the illness not been diagnosed at an early stage, there is a possibility the patient would not have survived. In this case, the pericarditis appeared to be triggered by endothelial cell injury with resultant thrombosis and supervening ischemic complications. There is limited literature on this topic.

TCTAP C-173

Anchor-monorail Technique Using Goose-neck Snare for Challenging Case in CRT Implantation

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[Clinical Information]

Patient initials or identifier number:

2822976

Relevant clinical history and physical exam:

A 56-year-old man without significant previous cardiovascular history was first admitted to another hospital with decompensated congestive heart failure manifested by significant orthopnea and lower extremity edema. His symptom significantly improved with using diuretics, also Amiodarone and Pimobendan added against non-sustained ventricular tachycardia and low output condition respectively. And then he was referred to our hospital for advanced treatment for heart failure, right heart catheterization and beta blocker titration. Patient had refused the CRT implantation because of anxiety associated with operative treatment back then. After 1 year later he was provoked syncope during walking, so re-admitted to our hospital for implantation of CRT with implantable cardioverter defibrillator (CRTD).

Relevant test results prior to catheterization:

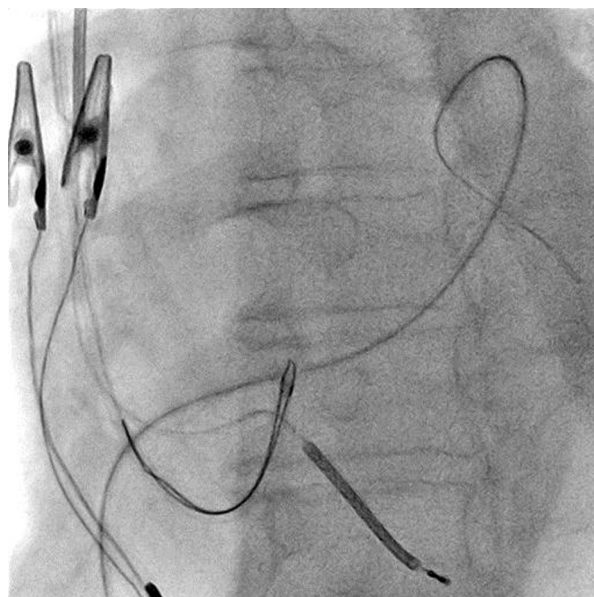
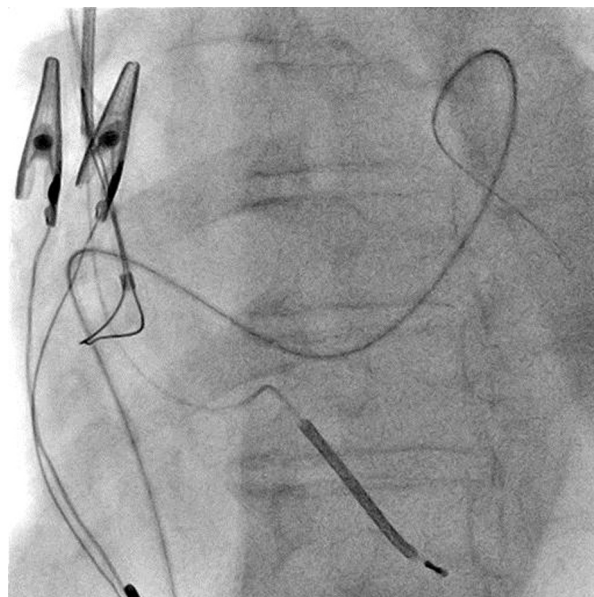
Electrocardiography showed wide PR and QRS duration, 242ms and 186ms respectively. Chest X ray shows marked cardiomegaly (CTR55%) and lung congestion. Transthoracic echocardiography showed markedly dilated LV (76mm), reduced LV ejection fraction (LVEF=26%), and moderate to severe mitral regurgitation.

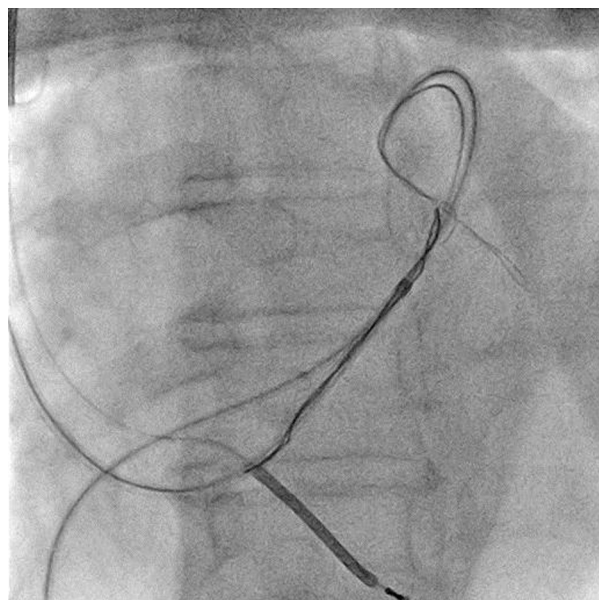
[Interventional Management]

Procedural step:

Blood access was gained via left subclavian vein using the Seldinger technique. We tried to cannulate Guiding catheter (GC) into Coronary sinus (CS) with a usual technique. But we could not get a CS ostium in spite of changing type of GC (CS-H; model 7075, CS-A6; model 7078 (Amplatz), Boston Scientific Japan) and using 0.035" guide wire (GW), inner catheter (CS-IC90) together with a fluoroscopic image. After right radial artery puncture was made and placed 4Fr Sheath, JL3.5

(4Fr, model JL-35B-SH, TORAY MEDICAL, Japan) was advanced to left coronary artery (LCA) ostium after injection of 2,000 u of Heparin. CS venography in delayed phase of LCA angiogram delineated the courses and ostium of the CS. The level of insertion of CS ostium located high above tricuspid annulus level. We thought it seems to get CS using 0.035" GW from femoral access because of significant disposition and tortuosity of proximal segment of CS ostium. If it was feasible, we could apply "the Anchor-Monorail technique" using the Balloon Angiographic with Endo Hole Catheter (BAC) as a rail after anchoring the BAC into distal CS. More specifically, the Goose Neck Snare (GNS) was advanced into RA through the GC via left subclavian access at first. The BAC was advanced into RA after placing 6Fr Sheath in right femoral vein on the next step. The BAC was anchored into distal CS after putting the BAC through a loop of GNS in RA. We tried to press the GC with GNS into CS with advancing GC with pulling back GNS making the tip of the GC to slide on the shaft of anchored BAC. In spite of every effort, we could not press the GNS into CS ostium. Eventually we could press the GC with GNS into CS after advancing 0.035" GW through GC into CS to make it coaxial the axis of CS ostium and distal tip of the GC with giving traction for the BAC anchored into CS simultaneously. A 0.014" GW (WHISPERVIEW™ EDS Guide Wire, model 4648, Boston Scientific Japan) was successfully advanced through GC (CS-H, model 7075, Boston Scientific Japan) and placed into anterior-lateral cardiac vein. The Unipolar IS-1 LV lead (ACUTY™ Spiral model 4592, Boston Scientific Japan) was advanced over the wire without any difficulty. Excellent pacing thresholds were obtained without any diaphragmatic stimuli. All coronary sinus GC were split and withdrawn and GW were removed. CRTD (INCEPT™, model P162, Boston Scientific Japan) was then implanted.





TCTAP C-174

Successful Treatment of a Pericardial Hemangioma with a Stent Graft

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[Clinical Information]

Patient initials or identifier number:

A 53 years old woman without systemic disease presented to the emergency department because of several episodes of atypical chest pain and palpitations.

Relevant clinical history and physical exam:

Her general physical examination showed decreased heart sounds.

Relevant test results prior to catheterization:

Her ECG, chest x-ray, and blood laboratory tests were unremarkable. Transthoracic echocardiography revealed a moderate amount of pericardial effusion and a hyper-echoic mass measuring 2.3 cm in length located to the left heart. Consequently, the patient was admitted to the cardiology intensive care unit for further evaluation. Advanced investigation of the mass was done using cardiac magnetic resonance imaging (MRI) and 256-slice computed tomography (CT). Cardiac MRI showed a moving ovoid-shaped and well-defined mass measuring 3.9×2.6 cm in the left pericardial space which was favored to be a benign tumor. CT of the chest, with intravenous contrast, disclosed a tumor with an ample blood supply along with a hemopericardium. This tumor was heterogeneous on unenhanced phase, mildly enhanced on arterial phase and intensely enhanced on venous phase after injection of contrast medium. Besides, two liver hemangiomas were found incidentally.

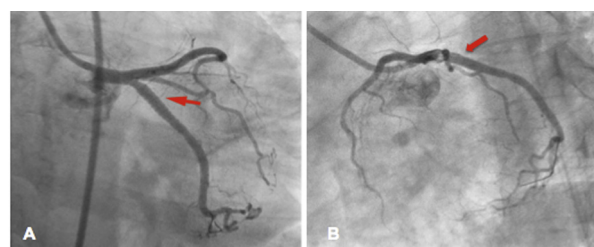
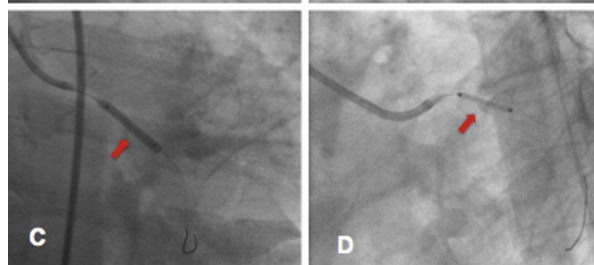
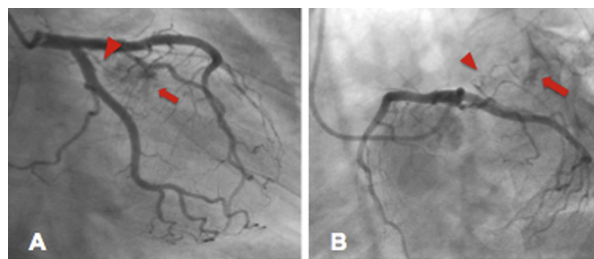
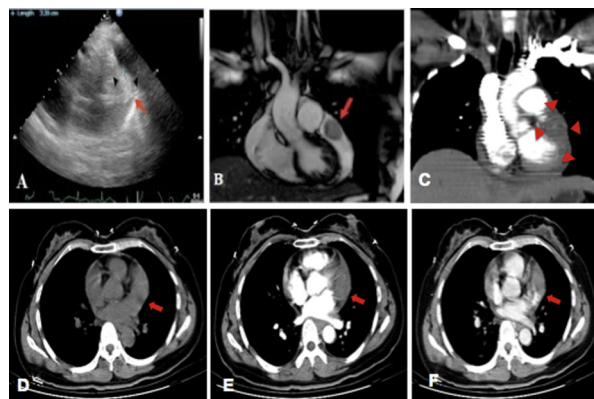
Relevant catheterization findings:

Coronary angiography revealed a small tortuous feeding artery arising from left circumflex artery and vascular lakes with "tumor blush" at the corresponding location.

[Interventional Management]

Procedural step:

Percutaneous coronary intervention was arranged. A 7-Fr JL4 guiding catheter was inserted into the left coronary artery, and a 0.014-inch floppy guide wire was placed into the distal left circumflex artery (LCX). The feeding artery of the cardiac hemangioma was too small to perform coil embolization. Therefore, a 3.5×19 -mm Jostent GraftMaster (coronary graft stent, Jomed, Helsingborg, Sweden) was advanced into the LCX without predilatation to close the orifice of the feeding artery. The stent graft was deployed at 14 atmospheres and was dilated using a 4.0×12 -mm balloon at 14 atmospheres to obliterate the coronary blood flow into the feeding artery of the cardiac hemangioma. After deployment of the stent graft, the patient received dual antiplatelet therapy with aspirin 100 mg and clopidogrel 75 mg per day for three months, after which the patient was maintained on clopidogrel 75mg per day.



Case Summary:

Cardiac hemangiomas are rare, non-malignant vascular tumors with diverse clinical presentations. The current treatment of cardiac hemangiomas depends on the clinical symptoms, and the primary treatment option is surgical resection. Coronary stent grafts have been used for treatment of coronary artery perforation, coronary pseudoaneurysm, and coronary artery fistula. In this article, we presented a 53 years old female patient with cardiac hemangioma complicated by pericardial effusion was treated with a stent graft. While surgical resection is the mainstay of treatment, coronary stent graft implantation may be an effective option for patients that need less invasive treatment for cardiac hemangiomas.